

Sarcomatoid Renal Cell Carcinoma: A Rare Case with Review of Literature

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Abstract

Case Report

Sarcomatoid renal cell carcinoma is a rare and highly lethal form of kidney tumor. It is characterised by extensive chromosomal rearrangements. These tumors show microscopic features similar to spindle cell sarcoma with high cellularity and cellular atypia. Majority of cases represent tumours with 100% sarcomatoid pattern and no recognizable epithelial element. We present a case of sarcomatoid renal cell carcinoma to highlight the clinical features and aggressive nature of this tumor.

Keywords: Renal Cell Carcinoma, Sarcomatoid, Aggressive.

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INTRODUCTION

Sarcomatoid renal cell carcinoma is a rare type of renal cell carcinoma which accounts for 1- 5% of renal cell carcinoma. This tumor is associated with worse prognosis [1, 2]. It represents a renal cell carcinoma with loss of epithelial phenotype and gain of mesenchymal features with extensive chromosomal rearrangements [3, 4].

CASE REPORT

A 16 year's female patient presented with pain in abdomen, hematuria since 2-3 months. CT abdomen

revealed large, well defined, soft tissue density lesion measuring 8.7 x 7.4 x 4.7 cm arising from upper pole of left kidney extending into para renal space with stag horn calculus measuring 4.9 x 3.7 x 2.2 cm in pelvis.

Left radical nephrectomy was performed and the specimen was sent for histopathological examination. Gross examination of left kidney revealed a tumor arising from upper pole, measuring 10.2 X 8.1 X 7 cm, cut section of which was grey white, fleshy with areas of haemorrhages and necrosis. The tumor was infiltrating perirenal fat (figure 1).



Fig-1: Gross image, Cut section of tumor is grey white, fleshy with areas of haemorrhages and necrosis

Microscopic examination revealed a tumor is arranged in diffuse fashion showing enlarged, highly pleomorphic and hyperchromatic nuclei with abnormal mitotic figures, many tumor giant cells and areas of haemorrhages and necrosis. Epithelial component was not seen with extensive sampling of tumor (figure 2 and 3). The tumor was infiltrating peri nephric fat. Five lymph nodes were isolated from perinephric fat out of which 2 lymph nodes revealed metastasis of sarcomatoid tumor (Figure 4 and 5). Multiple renal calculi with one stag horn calculus was noted, microscopy from corresponding renal tissue revealed features of chronic pyelonephritis.

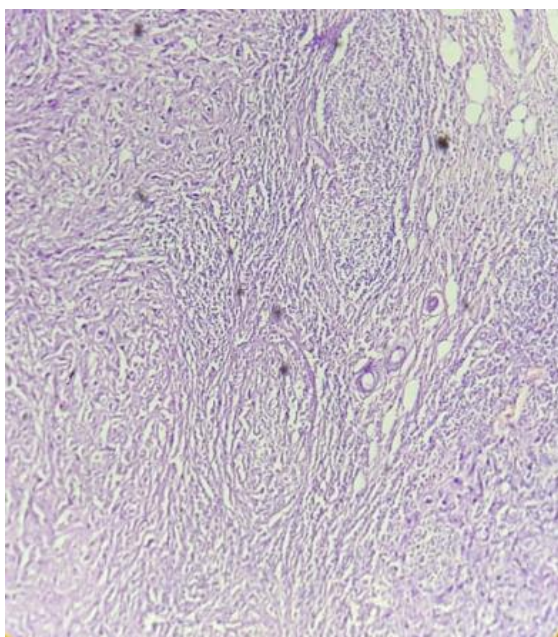


Fig-2: Tumor with spindle shaped neoplastic cells arranged in diffuse fashion, H and E stain, scanner view)

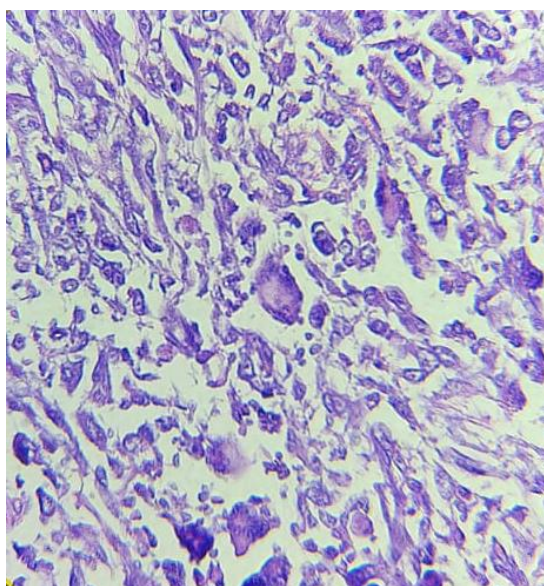


Fig-3: Tumor cells are enlarged, highly pleomorphic and hyperchromatic nuclei with abnormal mitotic figures, many tumor giant cells, H and E stain, 400X)

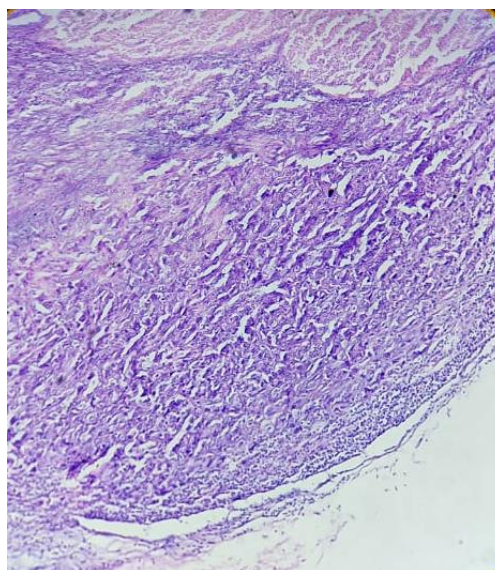


Fig-4: lymph nodes revealed metastasis of sarcomatoid tumor, H and E stain, scanner view)

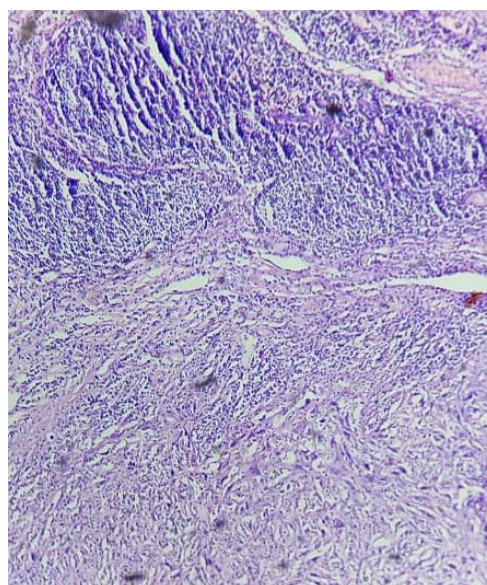


Fig-5: lymph nodes revealed metastasis of sarcomatoid tumor, H and E stain, 100X)

DISCUSSION

Sarcoma like histology in renal tumor is not a distinct entity but a pathway of transformation in renal cell carcinoma. Mean age of these patients is 60 years with male predominance. These patients present with gross hematuria, abdominal pain as observed in our case [5]. Presence of even a focal sarcomatoid component is associated with worse prognosis [1]. Sarcomatoid change can be uniform or heterogeneous. Heterogeneous pattern shows osteoid or chondroid differentiation. Additional high-risk factors are necrosis and micro vascular invasion [2]. Majority of these tumors have carcinoma elements [6]. Patients with chromophobe renal cell carcinoma reveal highest frequency of sarcomatoid transformation [7]. In our case, entire tumor revealed sarcomatoid component and

no epithelial component but metastasis in perinephric lymph nodes were noted.

Differential diagnosis of sarcomatoid renal cell carcinoma is primary renal cell sarcoma which is extremely rare in adults. Other differential diagnosis is sarcomatoid urothelial carcinoma.

Confirmation of sarcomatoid renal cell carcinoma may be done with additional tests like electron microscopy and immunochemistry. Immunohistochemistry for common epithelial and mesenchymal markers are helpful for differentiation. Sarcomatoid areas still express cytokeratin AE1/ AE3 [8]. Differential diagnosis of sarcomatoid renal cell carcinoma is primary renal sarcoma which accounts for less than 1 % of renal malignancy, in which areas of renal cell carcinoma are not seen. Sarcomatoid renal cell carcinoma is large tumors with high rate of metastasis and worst prognosis [9, 10, 1].

In a series of cases with nephrectomy more than 33% had positive lymph nodes as in our case [6]. Cytoreductive surgery has a significant role in positive advanced cases of sarcomatoid renal cell carcinoma, but survival benefits are questionable [11].

CONCLUSION

Sarcomatoid renal cell carcinoma is rare and represent terminally differentiated clone arising from conventional renal cell carcinoma. The tumor reveals aggressive course. Histopathological examination is the only method for definitive diagnosis.

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